Asymptomatic, primary prevention. Two patients (patients 13 and 18) in this cohort underwent VATS-LCSD as a prophylactic measure. The outcomes were equivocal as to whether any real benefit was derived from undergoing the procedure. With such limited experience and indeterminate results, our data do not support undertaking prophylactic VATS-LCSD in all children identified at high risk of fatal arrhythmias. Nevertheless, there may be a subset of patients who possess a high-risk genotype that would benefit from prophylactic LCSD treatment. A recent study by Jons and colleagues[21] demonstrated that analysis of mutant-specific ion channel characteristics in LQTS patients may be useful for clinical risk stratification. Further investigation may reveal a specific genotype and phenotype that are associated with a heightened risk profile and would, therefore, benefit from adjunctive VATS-LCSD as a primary preventative strategy. Nevertheless, we cannot recommend LCSD as a prophylactic therapy.

CONCLUSIONS

In summary, we have documented that video-assisted thoracoscopic LCSD can be safely and effectively performed in most children and young adults with life-threatening ventricular arrhythmias. This minimally invasive procedure is a promising adjunctive therapeutic option that achieves a beneficial response in most symptomatic patients. We advocate the use of this treatment in all patients who remain symptomatic with recurrent life-threatening arrhythmias, syncope, or frequent ICD discharges, despite conventional medical therapy. This treatment strategy should be considered as part of the treatment armamentarium in all patients with recalcitrant ventricular arrhythmias. The utility of LCSD as a prophylactic therapy in high-risk pediatric patients must be further elucidated before definitive recommendations can be made.

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Discussion

Dr Joseph Dearani *(Rochester, Minn)*. Thank you, Dr Backer and Dr Reddy.

Dr Hofferberth and colleagues have summarized their results of a small series of 24 children with VATS sympathetic denervation to treat life-threatening ventricular arrhythmias. Their technique intentionally spares the entire left stellate ganglion. They demonstrated a ''marked reduction'' of arrhythmias in 73% and elimination of arrhythmias in 55%. Two were lost to follow-up, and 2 had 1-month follow-up; this should be factored into the recurrence equation.

Surgery was performed safely; however, I believe there are shortcomings with this review. Most centers with the greatest experience in treating these cardiac channelopathies intentionally remove the lower half of the left stellate ganglion. The literature has demonstrated that the optimal cardiac denervation includes the removal of T4, T3, T2, and the lower pole of the left stellate ganglion (T1). In fact, the greatest density of norepinephrine-containing vesicles resides in the stellate ganglion (T1) and a portion of T2. So, ideally, cardiac denervation would include a complete

stellectomy. However, the upper pole of the left stellate ganglion is preserved to minimize the potential risk of developing Horner syndrome.

Among the largest programs that perform this specific left cardiac sympathetic denervation operation (ie, taking the lower half of the stellate ganglion and T2 through T4), there exceeds a 90% reduction in arrhythmia burden that includes breakthrough faints and breakthrough ICD shocks overall. Furthermore, the antifibrillatory (ie, protective effect) of denervation therapy is disease and disease genotype dependent, where it has been shown to be most effective in LQT1 and CPVT, emphasizing the importance of genotyping for all of these patients. For example, among the greater than 30 LQT1 patients denervated in our practice, there have been no breakthroughs to date with longer follow-up. Unfortunately, we do not know the genotypes of the long QT patients in this small series. If many or most were LQT1, then the higher observed breakthrough rate further underscores the critical importance of including the lower pole of the left stellate ganglion in the operation.

I believe cardiac denervation surgery for channelopathies should not be viewed as a simple adaptation of minimally invasive surgery performed for hyperhidrosis, and it probably should not be performed at a pace of approximately one procedure per year. Despite performing approximately 20 per year, we still encounter pretty variable anatomic variation with the left stellate ganglion. In addition, in our experience, we have now performed left cardiac sympathetic denervation in over 110 patients with these potentially life-threatening disorders. In the early part of our reported series, there were 3 patients with eyelid ptosis. Importantly, to date, there is no patient with a complete Horner (facial droop), in more than 110 patients.

In closing, I agree with your VATS approach to sympathectomy, and I congratulate your team for performing it with low periprocedural morbidity. However, I respectfully disagree with your technique to intentionally spare the left stellate ganglion, leaving up to a third of your patients with residual ventricular arrhythmias, resulting in ICD discharges. This study demonstrates that this technique is less effective for this difficult problem, and I would encourage you to acknowledge this higher incidence in arrhythmia recurrence when the lower half of the stellate ganglion is preserved.

Dr Hofferberth. Thank you very much for your comments, Dr Dearani.

First, I would like to address the issue of the indications for treatment in this study. The vast majority of the literature that exists using the VATS-LCSD procedure is in patients with cardiac ion channelopathies, who are treated for secondary prevention. Looking at our data, we had a 73% response rate among all-comers with 2 patients in the series of 24 treated for primary prevention. If you exclude those 2 patients out, that means that we have an 82% response rate among the patients that were treated for secondary prevention.

Published data from other centers that perform the VATS-LCSD technique certainly show that an 82% response rate is equivalent to the results attained across the entire global experience to date. There was a review article published 2 years ago out of Texas Children's Hospital by Hwang and colleagues, who reviewed all cases using VATS-LCSD procedure in children with congenital long QT syndrome. This article demonstrated that, of the global experience, there was a 77% response rate, defined in terms of arrhythmia reduction. The vast majority of data available on this topic defines response to treatment as reduced arrhythmia burden. As I alluded to in the presentation, it is difficult to quantify arrhythmia burden; however, that is a limitation across all of the studies in this area. Nevertheless, based on the current criteria for treatment response, our results are equal to the global experience.

Dr Dearani. There is an important difference between a 90% to 95% success rate, with ICD discharges going off in children with refractory ventricular arrhythmias compared to 55% to 70%. This is an evolving science. Genotyping has helped understand expectations in abolishing arrhythmias substantially. We have not had a single case of a permanent Horner syndrome with facial droop in over 100 cases when the lower pole of the stellate is removed. Life-threatening ventricular arrhythmia recurrence, resulting in ICD discharge, is different than recurrence of atrial fibrillation. It is a truly life-threatening problem, and striving for a 90% to 95% arrhythmia reduction rate without causing complete Horner syndrome seems to be a better goal to aim for.

It would be helpful for other surgeons in the audience to comment. As we discussed earlier, general pediatric surgeons and pediatric cardiac surgeons do this procedure. It is important that the surgical community understand the differences and expectations with the various sympathectomy techniques.

Dr Hofferberth. Thank you, Dr Dearani. Our institution does not agree with the notion that removal of part or all of the left stellate ganglion is a morbidity-free approach. The most recent article published out of the Mayo Clinic in 2012, which described their experience performing VATS-LCSD with resection of the lower half of the left stellate ganglion, demonstrates this is not a morbidity-free approach, with just over 10% of patients developing a perioperative Horner syndrome.

There was also an article published last month by Schneider and colleagues from a center in Munich, who reported their results performing LCSD with resection of the lower half of the left stellate ganglion in 10 patients with long QT syndrome. They reported that 7 of the 10 developed perioperative Horner syndrome. Our approach has been to transect the sympathetic chain up, at the base of the stellate ganglion, and in the process remove all of the interior radiating nerve fibers, which is, as you state, the location that releases the highest concentration of norepinephrine. By performing the procedure in this fashion, we are effectively eliminating the morbidity risks associated with this procedure without sacrificing the treatment efficacy.

Dr James Tweddell *(Milwaukee, Wis)*. Thank you. It was a nice presentation. And although I would agree with Dr Dearani that this is not a simple extension of a thoracoscopic sympathectomy for hyperhidrosis, it is probably closer to that than the operations typically performed by most of the people in this room. So, when we have encountered this problem, we have actually asked our adult thoracic surgery colleagues to help us with this procedure. They have tremendous experience with this. And, I think it is easier for us to work with them as a team approach rather than try to reinvent the wheel on these patients; I did enjoy your presentation much.

Dr Hofferberth. Thank you much for your comments, Dr Tweddell.

Dr Laureano Molins *(Barcelona, Spain)*. We have experience with sympathectomy or better clipping for hyperhidrosis mainly. We have little experience, 5 or 6 cases with babies. It is difficult to decide if the stellate ganglion should be removed or not because we do not want to have those Horner syndromes.

I think that the theory is to transect the lower part of the stellate ganglion, but it is not easy to respect the whole one. So, in fact, I have not done it, to resect the stellate ganglion, and the percentage of the patients that went well was similar, 75% to 80%.

But, I would like first to congratulate you for your elegant presentation and to talk a little bit about bilateral approach. Could this bilateral approach reach a high level of success? I really do not know. And we begin always with left side, of course. We go through until T6. But, I would like to know not only your experience but your opinion on that.

Dr Hofferberth. Thank you for your comments and question. At this point in time, the experience of performing a bilateral sympathectomy is limited at our center; however, the reason we did include the comment on the conclusion slide is that most recent patients treated with VATS-LCSD at our center had undergone a left-sided sympathectomy and remained symptomatic. In this particular case, we then decided to proceed with a second operation to perform a right-sided sympathectomy, and since that time, that patient has been completely arrhythmia free. It is, obviously, a limited experience to date. However, this may be a strategy that should be considered further in the future.

Dr Carl Backer *(Chicago, Ill)*. I have 2 questions. The first relates to a patient of ours with ventricular tachycardia treated by our chief of anesthesia with a temporary sympathetic nerve block in the left neck. This completely cleared up the arrhythmia and then we electively took the patient for a thoracoscopic sympathectomy several days later. Do you have any experience with temporary nerve blocks in the neck as a predictive study for this patient population?

Dr Hofferberth. As far as I am aware, there has not been any experience using that as a temporary measure.

Dr Backer. Yes, and it worked, it was unbelievable, it was night and day.

The second question I have relates to the fact that some of these children are pretty small and putting in an epicardial ACID is not without issues. Can we use sympathectomy as the primary therapy? We had one patient in whom we did the thoracoscopic sympathectomy, the arrhythmias went away, and we observed the child for a long time. We are still discussing whether or not we should put in an AICD. Do you have patients in whom you have simply done the sympathectomy and then not proceeded with an AICD?

Dr Hofferberth. There are certainly patients that have had a sympathectomy with additional medical treatment and who did receive an ICD. We view this procedure as an adjunctive therapeutic strategy that should always be implemented in conjunction with established therapies. In patients treated with VATS-LCSD for primary prevention, we would still treat them with optimal medical therapy; however, there have been some patients who have been spared from undergoing ICD implantation.

Dr Backer. I failed to mention we did treat this patient medically, and we did watch in the hospital for over a week. I noticed your mean hospital stay was 2 days. How long would you watch a patient with a sympathectomy and medical therapy before you would send them out?

Dr Hofferberth. We discharge all patients on medical therapy and then proceed to have them followed up regularly with a cardiologist. So, at this point, a patient that has received a sympathectomy is always going to be on some form of antiarrhythmic therapy.

Dr Backer. Thank you much. This really was an eye-opening presentation. This was not even on my radar screen 10 years ago, and certainly it has now become an effective therapy. Thank you much.